

primary studies - published RCT

Successful Therapy over 12 Months of People with Cystic Fibrosis with Rare Non-phe508del Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) Mutations with Elexacaftor/Tezacaftor/Ivacaftor (ETI).

Code: PM39727500

Year: 2024 Date: 1988

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Study design (if review, criteria of inclusion for studies)

The first study was an open randomized crossover trial. The second study consisted in a comparison of the response to Pancrease for at least 3 months (3-67 months) with the response to conventional pancreatin given to the same patients from 10 months to 8 years earlier.

Participants

children with cystic fibrosis (CF).

Interventions

1st study: enteric-coated microsphere preparation (Pancrease) versus conventional pancreatin given alone or with cimetidine. 2nd study: Pancrease for at least 3 months (3-67 months) with conventional pancreatin

Outcome measures

The parameters evaluated included the following: fecal fat excretion, coefficient of fat absorption, daily caloric intake, percent of diet as fats, proteins and carbohydrates, increase in height and weight, frequency and consistency of stools, palatability of the preparation, and patient compliance.

Main results

With Pancrease compared to conventional pancreatin, a significant improvement was observed in all the digestive parameters in addition to better patient compliance. In comparison to conventional pancreatin, Pancrease provided better digestive efficacy and greater increases in the growth rate of teenage patients. With Pancrease, the number of daily dosage units is decreased even when fat intake is increased. No adverse reactions were seen with either of the enzyme preparations used in these studies.

<http://dx.doi.org/10.3390/arm92060049>

See also

Adv Respir Med. 2024 Dec 20;92(6):559-572. doi: 10.3390/arm92060049.

Keywords

Adolescent; Child; Cimetidine; Combined Modality Therapy; Gastrointestinal Agents; Infant; Microspheres; pharmacological_intervention; Pancreatic Enzyme Replacement Therapy; Supplementation; Pancreas insufficiency; Pancreatic Diseases; Gastrointestinal Diseases; Malabsorption; Nutrition Disorders; Enteric-Coated; Histamine H2 Antagonists;